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Case Report

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A rare case of giant adrenal ganglioneuroma

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ABSTRACT

Ganglioneuromas (GNs) are rare benign tumors that typically occur in the posterior mediastinum or retroperitoneal region but can also be found in adrenal glands. Up to 50% of patients are asymptomatic, but symptoms can arise from the mass effect or excessive secretion of catecholamines. Careful evaluation is necessary to differentiate GNs from other adrenal tumors. A 19-year-old female patient with no previous comorbidities was diagnosed with a right adrenal incidentaloma. Physical examination and routine laboratory tests were normal. Imaging revealed a solid lesion in the right adrenal gland, and excisional adrenalectomy was performed to remove the mass, which was endocrinologically nonfunctional. Pathological examination confirmed the diagnosis of a right adrenal GN. The patient had an uneventful recovery and showed no recurrence during the follow-up period. After surgical removal of a benign neurogenic tumor, our patient had no tumor recurrence after five months, indicating a positive prognosis. However, long-term follow-up is still recommended because of reported cases of metastases and recurrence in some patients.

Keywords: Adrenal, ganglioneuroma, surgery, adrenalectomy

INTRODUCTION

Ganglioneuromas (GNs) are an uncommon benign tumor that develops from neural crest cells of the sympathetic nervous system.[1] GNs are most commonly found in the posterior mediastinum (38%), followed by the retroperitoneal region.[2] GNs are rarely found in adrenal glands (Figure 1).[3] Up to 50% of patients show no symptoms; most are incidentally detected. If patients develop symptoms, they are most commonly caused by the mass effect of the tumor. Infrequently, patients experience hypertension due to high levels of excessive secretion of catecholamines and their metabolism by the tumor.[4] According to previous reports, catecholamines or their metabolites are secreted by only 20–39% of GNs.[3,4] Additionally, because adrenal GNs can vary in size, imaging, and diagnostic characteristics, some of them might be mistaken for other adrenal tumors, such as adrenocortical carcinoma (ACC) and pheochromocytoma (PC).[5,9] Therefore, careful evaluation using endocrine examinations and multiple imaging procedures is required to rule out other types of tumors.[10]

In this report, we present a case of 19 years old female patient with a giant adrenal tumor incidentally diagnosed as adrenal GNs.

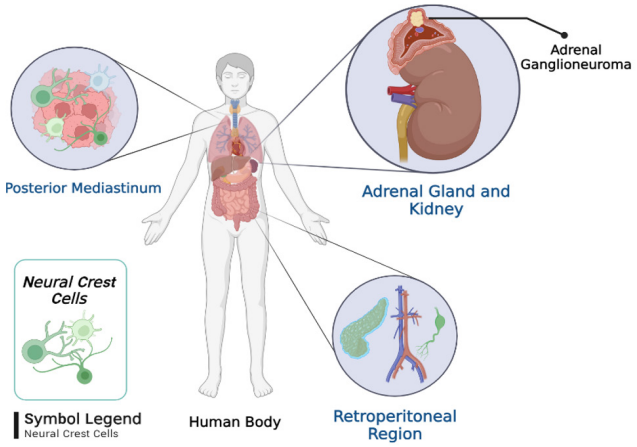

Figure 1. Ganglioneuroma (GN) is a rare benign tumor that originates from neural crest cells of the sympathetic nervous system. It is most commonly found in the posterior mediastinum (38%) and retroperitoneal location, while rarely found in adrenal glands

CASE

A 19-year-old female patient with no previous comorbidities was referred to our hospital after the discovery of a right adrenal incidentaloma during a workup for vague abdominal pain. On admission, she complained of vague

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abdominal pain, with no other complaints. In her past medical history, the patient denied comorbidities as well as signs and symptoms of hypercortisolism and virilization, high blood pressure levels, or paroxysms of headache, palpitations, or sweating. There was no history of smoking, drinking, recent travel, or relevant family history. There were no previous hospital admissions or surgeries in her medical history.

Physical examination revealed no signs, and the results of routine laboratory tests were all within normal ranges. Radiography and computed tomography (CT) of the thorax, as well as electrocardiography, were standard. The results of routine laboratory and urine tests and levels of malignant tumor markers, such as carcinoembryonic antigen (CEA) and CA19-9, were all within the normal ranges. Endocrine examination values included the following: standard adrenal plasma aldosterone/plasma renin activity ratio, normal androgen hormones, normal 24-hour urinary native catecholamines and metanephrines, normal adrenal corticotrophin and cortisol rhythms, and normal 24-hour urinary free cortisol. Adrenal corticotrophin stimulation test revealed a normal plane.

Abdominal CT showed a solid lesion in the right adrenal gland, approximately 9x6.5x14 cm in size, hypodense heterogeneous density with minimal contrast, and enlargement of the liver parenchyma. Magnetic resonance imaging (MRI) findings were unfavorable for the diagnosis of cortical adenomas (Figure 2). The patient underwent excisional adrenalectomy.

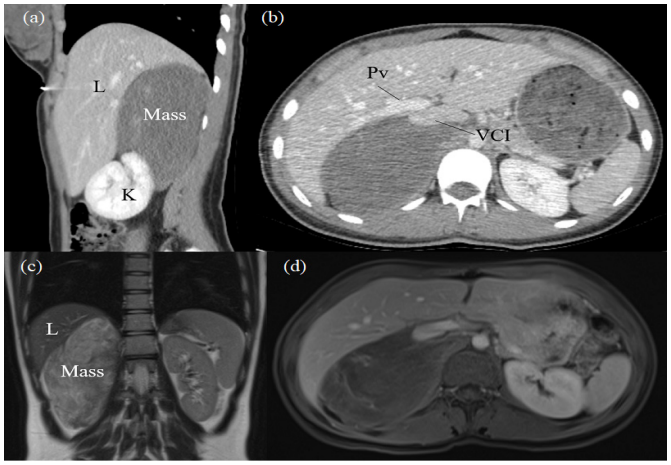

Figure 2. (a) and (b). The multislice CT shows a hypodense heterogeneous solid lesion with hypodense heterogeneous density and minimal contrast enhancement approximately 9x6.5x14 cm in size at the level of the right upper pole of the liver parenchyma, which is not easily distinguished from the right surrenal gland. On MRI images of the lesion, hyperintensities on T1-weighted images (c) and heterogeneous hyperintensities on T2 weighted images (d) are evident, causing contrast enhancement in the septations after intravenous contrast injection, and significantly compressing the right kidney and liver. L; liver, K; kidney, Pv; portal vein, VCI; vena cava inferior

The surgical specimen was an elastic hard tumor with a slightly lobular edge, measuring 15x11,7x6 cm (Figure 3). The cut surface of the tumor was light brown and covered with a thin capsule without any evidence of hemorrhage or necrosis. There was no evidence of malignancy. On pathological examination, the tumor was diagnosed as a right adrenal GNs (Figure 4). The patient was discharged without incident on postoperative day five. No recurrence was observed during the 5-month follow-up period.

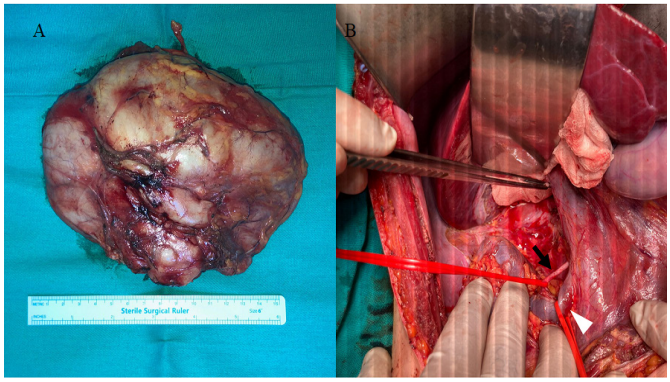

Figure 3. A. Macroscopic view of the specimen at the anatomical position. The right adrenal gland in the specimen weighed 570 g and measured 15x12.3x6 cm. It was a solid, homogeneous tumor with a grayish-white surface, occasionally covered by a thin transparent capsule, and sometimes with attached adipose and membranous tissue. B. Surgical field after specimen removal. A black arrow indicates the right renal artery, while a white arrowhead indicates the right renal vein

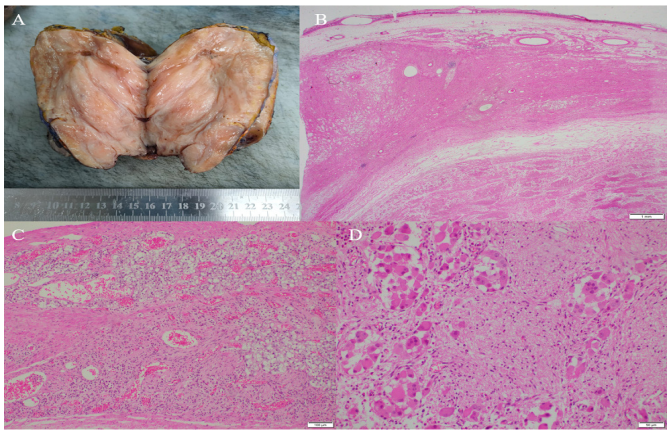

Figure 4. A. Mass of the adrenal gland, 15 cm in diameter, well-circumscribed, multilobulated, homogeneous off-white in color. On the inferolateral edge of the mass, the residual adrenal cortex is visible. B. Ganglioneuroma consisting of mature Schwann cells forming fascicles separated from the surrounding tissue by smooth borders (H&E, x12.5). C. Adrenal parenchyma (top) and ganglioneuroma (bottom) (H&E, x100). D. Mature spindle Schwan cells with large eosinophilic cytoplasm, eccentric nuclei, and prominent nucleoli, along with some multinuclear dysmorphic ganglion cells (H&E, x200)

DISCUSSION

In this report, we present the case of a 19-year-old female patient with giant adrenal GNs who was differentially diagnosed with ACC and PC.

The neural crest tissue of the sympathetic nervous system leads to a rare, benign, and slow-growing tumor known as GNs, which is histologically composed of mature Schwann cells and ganglion cells with fibrous stroma.[1] Additionally, because adrenal GNs vary in size, imaging, and diagnostic characteristics, some might be mistaken for other adrenal tumors, such as ACC and PC.[5,9] However, there are a large number of diagnostic differentials of an adrenal mass comprising a long list, including adenoma, myelolipoma, cyst, lipoma, pheochromocytoma, adrenal cancer, metastatic cancer, hyperplasia, and tuberculosis.[6,7,11-13] We retrospectively reviewed the hormonal status and CT and MRI features and compared this information with the histopathological findings to prevent misdiagnosis.

Adrenal incidentalomas are rare in patients under the age of 30 years but increase in frequency with age.[13] It affects both sexes equally, and the fourth and fifth decades are the most common.[14] Moreover, the pathophysiology of GNs is

influenced by genetic factors, including mutations in the tyrosine kinase receptor ERBB3.[14] In the present case, the patient's age (19 years) was not within the range mentioned in previous reports, and there was no family history of Adrenal GNs.

The posterior mediastinum, retroperitoneum, and adrenal gland are the primary locations of GNs. The adrenal medulla is where 20% of GNs are found.[2,3] As a result, GNs is rarely considered in the diagnosis of adrenal tumors. However, as seen in the present case, imaging techniques such as ultrasonography (US) and CT has recently expanded, leading to an increase in the number of adrenal GNs found incidentally. It is estimated that an adrenal tumor is incidentally found in 1-10% of cases of abdominal CT, and 1-6% of these are GNs.[6,8,15,16] Adrenal GNs have been characterized as well-circumscribed tumors with an oval or lobular form, minimal attenuation, and homogenous or moderately heterogeneous masses in terms of imaging properties on CT.[9] On MRI, adrenal GNs shows homogenously low or intermediate signal intensity on T1-weighted images and heterogeneous slightly high signal intensity on T2-weighted images.[17] The CT and MRI imaging characteristics were consistent with those of our case.

Hormonally silent adrenal GNs are typically found incidentally. The most frequently reported secondary symptom, as in our case, is back and abdominal pain caused by the mass effect of the tumor.[1,3] The patient in this report had an incidentaloma, which is the most common manifestation of adrenal GNs. The patient may develop particular symptoms, including hypertension, diarrhea, weakness, and virilization when there is an excessive release of catecholamines or steroid hormones (indicative of a hormonally active disease).[4] However, these symptoms were not observed in our patient. Therefore, surgical excision is required to confirm this diagnosis.

Surgery is the initial treatment option for adrenal GNs, particularly for large tumors. According to the National Institutes of Health State-of-the-Science Statement, non-secretory adrenal incidentalomas with dimensions >6 cm or suspicious features of malignancy should undergo adrenalectomy because of the high prevalence of malignancy in non-secretory adrenal incidentalomas.[18] In the present case, the tumor size was >12 cm; therefore, we performed excisional tumor resection. As laparoscopy is less invasive than standard open surgery, it may be a superior surrogate. However, because the tumor was large and the potential of adrenal cortical carcinoma could not be ruled out in our case, we decided to perform an open tumor excision. The likelihood of a malignant tumor is higher among lesions larger than 4.5 cm, even though histological analysis may not confirm it. While our patient's tumor measured >12 cm in diameter, the pathology report indicated that it was benign. Typically, adrenal GNs consist of encapsulated masses with a solid, grayish-white cut surface, are homogenous in structure, as seen in the present case, and often have a firm consistency.[10]

CONCLUSION

Our patient showed no signs of tumor recurrence five months after surgery. Although there have been a few reported cases of metastases and recurrence, an excellent prognosis exists for patients who have undergone surgical removal of a benign neurogenic tumor without any further treatment requirements. Therefore, a long-term follow-up is recommended after surgery.

ETHICAL DECLARATIONS

Informed Consent: The patient signed the free and informed consent form.

Referee Evaluation Process: Externally peer-reviewed.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

Financial Disclosure: A conflict of interest has not been declared by the author.

Author Contributions: All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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I am a fifth-year medical student at Kahramanmaraş Sütçü İmam University. Ever since I was young, I always loved human science. I just enjoyed everything about it. How the body functions interests me, both in health and illness, how to find out what was wrong and how to address it. It has always been my dream to be a doctor. I appreciate Gazi University and Prof. Dr. Mehmet Akif Türkoğlu for this opportunity of scientific research and I'm looking forward to contribute in other studies also.